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The scimitar syndrome: About two observations.

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Abstract: *We report two cases of scimitar syndrome in two infants, aged of 6 and 14 months. The circumstances of the discovery were dyspnea, fever, fatigue at feeding time, and respiratory distress. The thoracic CT angiography is a non-invasive examination, which has enabled us to recognize scimitar syndrome, identify anomalies of systematization of the right lung, to show the anatomical presence of the right pulmonary artery, and to determine whether it has been opacified normally in the anterograde direction at the pulmonary arterial time, or abnormally in the retrograde direction at the systemic arterial time, from a systemic artery of the abdominal aorta.*

Keywords: scimitar syndrome, abnormal pulmonary venous return.

Introduction

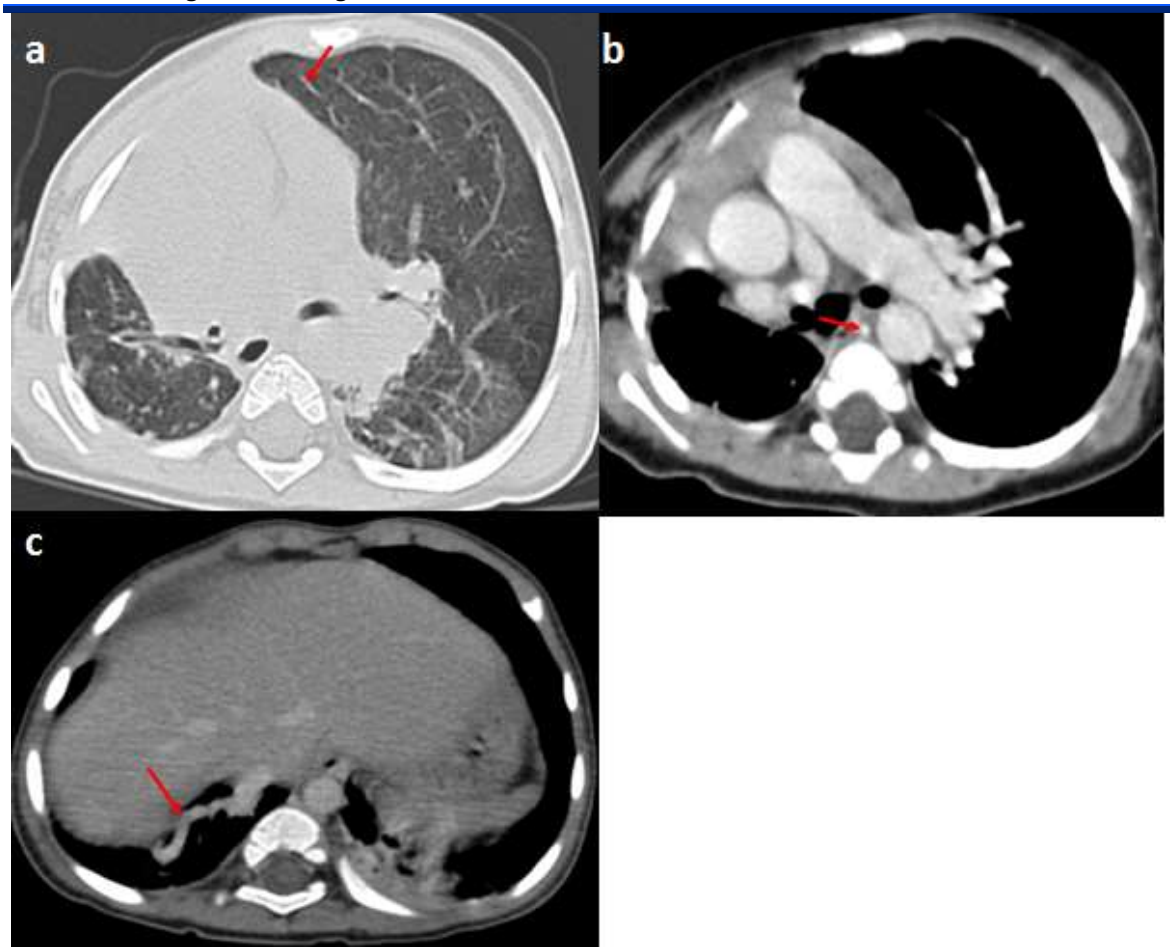
The scimitar syndrome associates a right partial abnormal pulmonary venous return in the inferior vena cava, generally immediately under the diaphragm, a hypoplasia of the right parenchyma and pulmonary artery, and sometimes a right basal sequestration vascularized by arteries coming from the abdominal aorta. This malformation occurs in 1 to 3 per 100,000 births / year with a sex ratio of 2/1 [1]. The clinical expression is very variable, ranging from intolerance in the first days of life to a fortuitous finding in adults; it depends on the importance of pulmonary hypoplasia and arteriovenous fistula through sequestration.

Observations:

Case 1:

We report the case of a 14-month-old infant admitted to emergency departments for dyspnea, fever and fatigue at feeding time. The clinical examination notes a right hemithorax less developed than the left. He was put under symptomatic treatment of bronchiolitis, with persistent of the dietary difficulties. Radiography of the thorax showed parenchymal distension of the left lung, and cardiomegaly with cardiac dextroposition. The echocardiography found a right partial abnormal pulmonary venous return in the right atrium, a pulmonary arterial hypertension (PAH) and signs of high blood flow. The CT angiography showed hypoplasia of the right lung, without individualization of the upper lobe (aplasia of the right upper lobe), with anterior mediastinal hernia of the left lung [Figure 1a]. The trunk of the pulmonary artery measured at 17 mm transverse diameter with a very hypoplastic right pulmonary artery measuring 06 mm and a left pulmonary artery measuring 12 mm [Figure 1b]. Moreover, absence of visualization of the upper right pulmonary artery, the right lower pulmonary artery is present but very hypoplastic. Cardiomegaly was also noted at the expense of the right cavities with dextroversion. There is an abnormality of the right partial pulmonary venous return with a lower right pulmonary vein which flows into the supra-diaphragmatic VCI (Figure 1c). The systemic vascularization for the right lung came from the celiac trunk, whose caliber is increased measuring 08 mm in diameter. All the radiological abnormalities allowed the diagnosis of the scimitar syndrome.

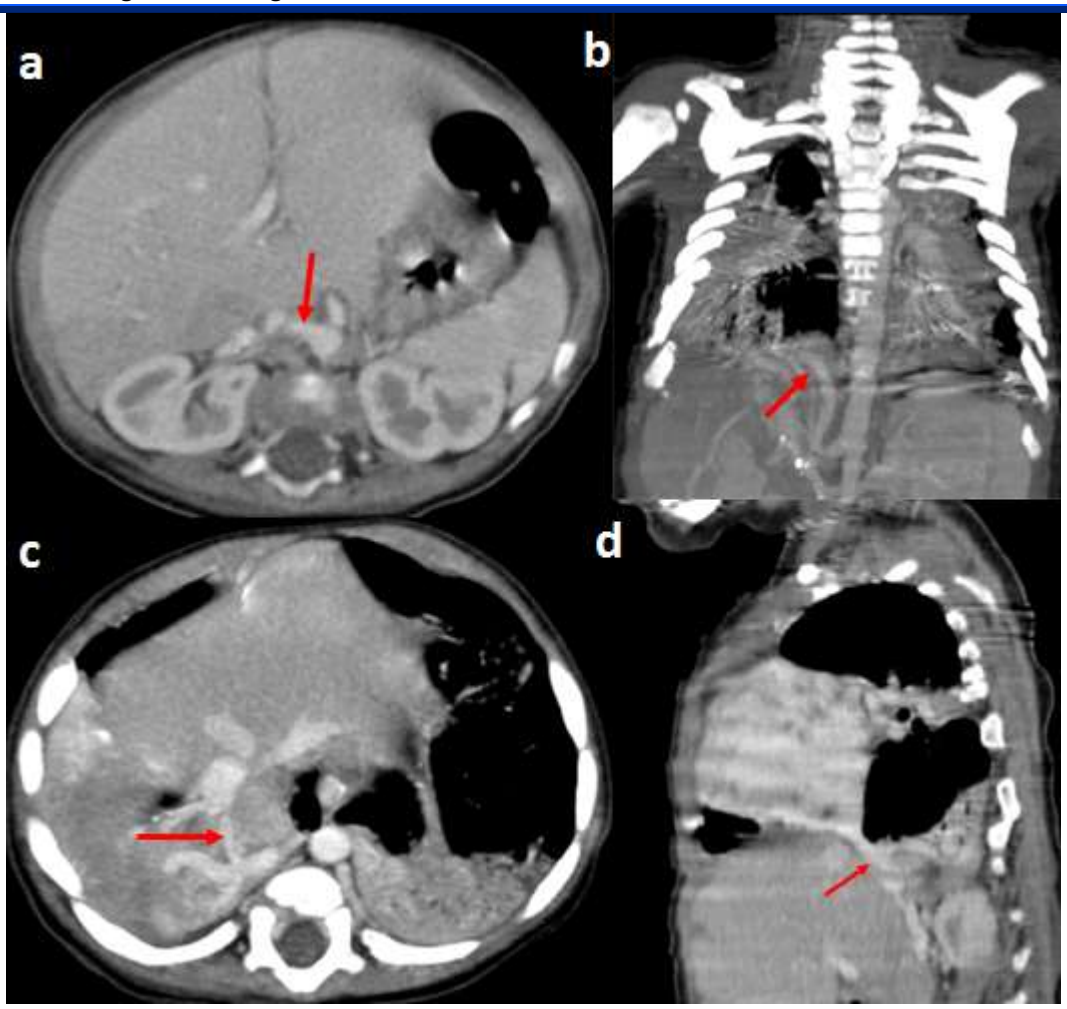
Figure 1



Case 2:

Infant of 6 months, admitted to emergency for respiratory distress. The clinical examination found a febrile and polypneic child with supra-sternal retraction, asynchronous chest and abdominal wall motion, and labial cyanosis. On the chest x-ray, there was an important chest distention (emphysematous left lung), and diffuse alveolo-interstitial syndrome. The patient was put under antibiotic and corticosteroid therapy without improvement. Echocardiography showed cardiac dextroposition, right ventricular dilatation, and PAH. CT angiography revealed an artery originating from the abdominal aorta [Figure 2 (a, b, d)], vascularizing a lower right pulmonary sequestration, with abnormal pulmonary venous return in the inferior vena cava [Figure 2c], which asserts the diagnosis of the scimitar syndrome.

Figure 2



Axial sectional scan (a), coronal (b), and sagittal reconstruction (d) in arterial time, showing the emergence and pathway of the systemic artery, which vascularizes right basal lung sequestration.

Axial sectional scan, at arterial time (c), showing the drainage vein of pulmonary sequestration at the level of the inferior vena cava.

Discussion:

Scimitar syndrome (congenital veinolobar syndrome or hypogenetic right pulmonary syndrome) is characterized by an abnormal pulmonary venous return draining into the inferior vena cava. The right pulmonary vein usually has a curved aspect in inside, hence the name of scimitar syndrome [2]. It is rarely associated with cardiac abnormalities, but is often associated with abnormalities of the right lung, its bronchial systematisation and vascularization (right pulmonary hypoplasia of variable degree), dextrocardia, and systemic vascularization of the base of the right lung by Abnormal arteries coming from the abdominal aorta (pulmonary sequestration). A competition between the pulmonary circulation which comes from the right heart and the systemic circulation which can be very important, may be at the origin of an inversion of the flow in the pulmonary artery, and thus of an arterio- Venous shunt between the systemic arteries and abnormal pulmonary venous return. The right pulmonary artery is often of small caliber as in our first observation, which reduces the left-right shunt of the abnormal venous return.

There are two main clinical forms [2]:

- **An infantile form:** (the case of our two patients) the right pulmonary artery is of small caliber, with little important a pulmonary artery-pulmonary vein-vena cava inferior shunt, but with a very important systemic vascularization coming from the Abdominal aorta, that determines a sometimes severe left-right shunt with severe pulmonary arterial hypertension and early cardiac insufficiency. The treatment consists on an embolisation or ligatures of abnormal systematic vessels.
- **Pediatric or adult form:** without pulmonary arterial hypertension, well tolerated. Pulmonary circulation is a little disturbed. Pulmonary arterial blood is drained by the pulmonary veins into the inferior vena cava, but the right pulmonary arterial circulation is weaker than the left and the shunt performed is generally small. Surgical replacement of the right pulmonary vein on the left atrium is debatable, given the risk of post-operative thrombosis of the anastomosis, which is complicated by recurrent haemoptysis and PAH, requiring sometimes right pneumonectomy. Sometimes pulmonary sequestration is associated, but without major repercussions.

A stenosis at the junction of the right pulmonary vein at the inferior vena cava has been described [3]. It creates an obstacle to right pulmonary venous return, causing PAH, if it is tight.

In some cases, the right pulmonary artery is absent, and the right lung is vascularized by systemic vessels. In others, the right pulmonary artery is hypoplastic, and the pulmonary circulation is countercurrent from the systemic vessels, the blood passes into the right pulmonary artery and then goes into the trunk of the pulmonary artery, and can even reach the left pulmonary artery.

Thoracic angiography is a non-invasive examination, which has allowed us to recognize scimitar syndrome, identify abnormalities in right lung systematization, show the anatomical presence of the right pulmonary artery, and whether It has been opacified normally in the anterograde direction at the pulmonary arterial time, or abnormally in the retrograde direction at the systemic arterial time, from a systemic artery of the abdominal aorta.

Conclusion:

Scimitar syndrome is a particular form of partial abnormal pulmonary venous return. It is a rare and unrecognized pathology, its clinical expression is insidious and unspecific. Its most severe form with pulmonary sequestration leads rapidly to a table of high-rate cardiac insufficiency whose management is a therapeutic emergency.

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